CENTERS FOR DISEASE CONTROL AND PREVENTION

September 15-16, 2010
Diagnosis Agenda, Part 2 of 2

Diagnosis Topics:

Disorders Due to Intrinsic Circulating Anticoagulants, Antibodies, or Inhibitors

The ICD-9-CM code 286.5, Hemorrhagic disorder due to intrinsic circulating anticoagulants, includes a number of diverse disorders, which are characterized by an abnormally elevated activated partial thromboplastin time (aPTT), despite different physiology.

One disorder is hyperheparinemia. This is an iatrogenic disorder, related to elevated heparin levels, from being given too much heparin. There are different types of heparin, including unfractionated heparin, and low molecular weight heparin. Both anti-IIa and anti Xa are specific types of purified heparin that are currently available.

Acquired hemophilia, or secondary hemophilia, is a disorder in which antibodies to a coagulation factor develop, usually coagulation factor VIII. This is also called autoimmune hemophilia. A specific code for acquired hemophilia will enable studies of new treatments for this disorder, that would otherwise be more difficult or impossible.

Systemic lupus erythematosus (SLE) inhibitor or lupus anticoagulant is an antibody directed against protein phospholipid complexes. There are certain other antiphospholipid antibodies that when present are a risk factor for thromboembolic disease, but patients may also be asymptomatic (e.g., anticardiolipin antibodies). The elevated aPTT is primarily related to interaction of antibody, such as lupus anticoagulant, with test reagents. While these antibodies do not typically cause hemorrhagic disease, there are some reported cases where such antibodies appeared to be related to bleeding. This would ordinarily require some other problem with blood clotting, although some cases have occurred where other problems were not found. When a hypercoagulable state is present, code 289.81 should be used. Presence of antibody without any diagnosis may be best coded to 795.79, Other and unspecified nonspecific immunological findings.

In order to better track the different diseases included at this code, it was requested that the code be expanded. This request was received from Novo Nordisk.

TABULAR MODIFICATIONS

286 Coagulation defects

286.5 Hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors

Delete

~~Antithrombinemia~~
~~Antithromboplastinemia~~
~~Antithromboplastino-genemia~~
~~Hyperheparinemia~~
~~Increase in:~~
~~anti-VIIIa~~
~~anti-IXa~~
~~anti-Xa~~
~~anti-XIa~~
~~antithrombin~~
~~Secondary hemophilia~~
~~Systemic lupus erythematosus [SLE] inhibitor~~

New code 286.51 Disorders due to iatrogenic anticoagulants
Hyperheparinemia
Anti-Xa
Anti-IIa

New code 286.52 Acquired hemophilia
Autoimmune hemophilia
Autoimmune inhibitors to clotting factors
Secondary hemophilia

New code 286.53 Antiphospholipid antibody with hemorrhagic disorder
Lupus anticoagulant (LAC) with hemorrhagic disorder
Systemic lupus erythematosus [SLE] inhibitor with hemorrhagic disorder
Excludes: Anti-phospholipid antibody, finding without diagnosis (795.79)
Anti-phospholipid antibody syndrome (289.81)
Anti-phospholipid antibody with hypercoagulable state (289.81)
Lupus anticoagulant (LAC) finding without diagnosis (795.79)
Lupus anticoagulant (LAC) with hypercoagulable state (289.81)
Systemic lupus erythematosus [SLE] inhibitor finding without diagnosis (795.79)
Systemic lupus erythematosus [SLE] inhibitor with hypercoagulable state (289.81)

New code 286.59 Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
Antithrombinemia
Antithromboplastinemia
Antithromboplastinogenemia
Increase in:
anti-VIIIa
anti-IXa
anti-Xa
anti-Xla
anti-IIa (thrombin)
anti-II (prothrombin)

289 Other diseases of blood and blood-forming organs

289.8 Other specified diseases of blood and blood-forming organs

289.81 Primary hypercoagulable state

Add Antiphospholipid antibody syndrome

Revise Lupus anticoagulant with hypercoagulable state

Add Systemic lupus erythematosus [SLE] inhibitor with hypercoagulable state

Excludes: Anti-phospholipid antibody, finding without diagnosis (795.79)

Anti-phospholipid antibody with hemorrhagic disorder (286.53)

Lupus anticoagulant (LAC) finding without diagnosis (795.79)

Lupus anticoagulant (LAC) with hemorrhagic disorder (286.53)

Systemic lupus erythematosus [SLE] inhibitor finding without diagnosis (795.79)

Systemic lupus erythematosus [SLE] inhibitor with hemorrhagic disorder (286.53)

INDEX MODIFICATIONS

Add Antibody
- anticardiolipin 795.79
  - with
    - hemorrhagic disorder 286.53
    - hypercoagulable state 289.81
- antiphosphatidylinositol 795.79
  - with
    - hemorrhagic disorder 286.53
    - hypercoagulable state 289.81
- antiphosphatidylglycerol 795.79
  - with
    - hemorrhagic disorder 286.53
    - hypercoagulable state 289.81
- antiphosphatidylserine 795.79
  - with
    - hemorrhagic disorder 286.53
    - hypercoagulable state 289.81
- antiphospholipid 795.79
  - with
    - hemorrhagic disorder 286.53
    - hypercoagulable state 289.81

Findings, (abnormal), without diagnosis (examination) (laboratory test) 796.4

Add antiphosphatidylinositol antibody 795.79

Add antiphosphatidylglycerol antibody 795.79

Add antiphosphatidylserine antibody 795.79

Interstitial Lung Diseases of Childhood

Neuroendocrine cell hyperplasia of infancy (NEHI) is an interstitial lung disease that occurs in children. NEHI is associated with prolonged oxygen use for years in children, starting in the first year of life and persisting with mild symptoms into adolescence. This disorder is increasingly being diagnosed, with published reports of infants with characteristic high resolution CT scan and pulmonary function test findings. Many children with NEHI undergo lung biopsies to make a final diagnosis, which is based on histological staining of neuroendocrine cells. Further emphasizing the importance of a diagnostic code for this disorder, familial cases are now being reported, suggesting that this disorder has genetic origins.

Neuroendocrine cell hyperplasia of infancy (NEHI) is not indexed in ICD-9-CM. The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for NEHI, as well as for a number of other interstitial lung diseases.

Pulmonary interstitial glycogenosis is a disorder increasingly recognized to complicate the course of newborns with out of proportion hypoxemia. Pulmonary interstitial glycogenosis results from the proliferation of a poorly defined clear cell population that contains glycogen in the alveolar interstitium, resulting in significant thickening of this space and marked diffusion abnormalities for oxygen. This disorder can occur as the primary finding or in associations with premature lung disease or congenital heart disease. It has major implications for children in the neonatal and cardiac intensive care unit. A lung biopsy is required to establish the diagnosis.

Pulmonary interstitial glycogenosis is not indexed in ICD-9-CM. It is not related to the glycogen storage diseases that are coded to code 271.0, Glycogenosis. The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for pulmonary interstitial glycogenosis.

The **surfactant mutations of the lung** are a group of disorders that cause significant morbidity and mortality for children and are a leading indication for pediatric lung transplantation. Children with these disorders most commonly present in the newborn period but may present in later childhood with unknown chronic lung disease. Each surfactant mutation has a characteristic clinical presentation, course and prognosis, and ideally each would benefit from separate ICD-9-CM codes. There is also growing evidence that these mutations may also be important gene modifiers for other lung disease, because of the important role surfactant plays in lung homeostasis. Specific ICD-9-CM codes would have an important role in identifying and tracking these genetic conditions.

The surfactant mutations include surfactant protein B mutation of the lung, surfactant protein C mutation of the lung, surfactant associated ATP binding cassette A3 mutation of the lung, and surfactant associated thyroid transcription factor 1 mutations of the lung (SPB, SPC, ABCA3, and TTF-1). The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of specific codes for the surfactant mutations of the lung.

Alveolar capillary dysplasia with vein misalignment (ACDMPV) is a developmental lung disorder. Children with ACDMPV present in the immediate neonatal period with rapidly progressive respiratory failure and severe pulmonary hypertension that progresses to death in the first 2 months of life despite therapeutic interventions for pulmonary hypertension, advanced ventilation strategies, and extracorporeal membrane oxygenation (ECMO). There are cases with a familial history suggestive of an

autosomal recessive genetic etiology. This is still an under recognized disorder diagnosed by lung biopsy or post mortem evaluation. The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for ACDMPV.

TABULAR MODIFICATIONS

Option 1 This would group surfactant mutations of the lung.

516 Other alveolar and parietoalveolar pneumonopathy

New subcategory	516.6	Interstitial lung diseases of childhood	
New code		516.61	Neuroendocrine cell hyperplasia of infancy
New code		516.62	Pulmonary interstitial glycogenosis
New code		516.63	Surfactant mutations of the lung
New code		516.64	Alveolar capillary dysplasia with vein misalignment
New code		516.69	Other interstitial lung diseases of childhood

Option 2. This would better specify each surfactant mutation of the lung.

516 Other alveolar and parietoalveolar pneumonopathy

New subcategory	516.6	Interstitial lung diseases of childhood	
New code		516.61	Neuroendocrine cell hyperplasia of infancy
New code		516.62	Pulmonary interstitial glycogenosis
New code		516.63	Surfactant protein B mutation of the lung
New code		516.64	Surfactant protein C mutation of the lung
New code		516.65	Surfactant associated ATP binding cassette A3 mutation of the lung
New code		516.66	Surfactant associated thyroid transcription factor 1 mutations of the lung
New code		516.67	Alveolar capillary dysplasia with vein misalignment
New code		516.69	Other interstitial lung diseases of childhood

Idiopathic Pulmonary Fibrosis

Idiopathic pulmonary fibrosis (IPF) is a distinctive type of chronic fibrosing interstitial pneumonia of unknown cause which is limited to the lungs. Diagnosis of IPF requires a number of findings, and exclusions. Surgical biopsy shows a 'usual interstitial pneumonia' pattern. Other known causes of interstitial lung disease must be excluded, including drug toxicities, environmental exposures, and collagen vascular diseases. Characteristic abnormalities are found on conventional chest x-rays or high-resolution computed tomography (HRCT) scans. Pulmonary function studies generally show restriction, with reduced total lung capacity or reduced vital capacity. There may be impaired gas exchange. Onset of symptoms is usually gradual, with dyspnea the most prominent and disabling symptom. A nonproductive cough is usual and may be paroxysmal, often refractory to anti-tussive agents. The patient's age at onset is usually greater than 50 years and IPF is slightly more common in males. The histologic pattern generally present with IPF is described by pathologists as usual interstitial pneumonia.

There is no specific ICD-9-CM code for IPF. It is also called cryptogenic fibrosing alveolitis, which is indexed in ICD-9-CM to 516.3, Idiopathic fibrosing alveolitis. However, that term is not typically used to describe IPF, and the code also includes Hamman-Rich syndrome, which is actually a form of acute interstitial pneumonia. The course of IPF is quite different from other interstitial pneumonias, including nonspecific interstitial pneumonia, the disease that it is most commonly confused with. A specific code will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for IPF, as well as for a number of other interstitial lung diseases. IPF is one of the idiopathic interstitial pneumonias, a group of scarring lung diseases with distinctive presentations, pathophysiology and clinical course.

References

Raghu G, Weycker D, Edelsberg J, Bradford WZ, Oster G. Incidence and prevalence of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med. 2006 Oct 1;174(7):810-6. Epub 2006 Jun 29. PubMed PMID: 16809633. (Available here: http://ajrccm.atsjournals.org/cgi/content/short/174/7/810).

"American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias"
http://ajrccm.atsjournals.org/cgi/content/full/165/2/277.

TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise	516.3	Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia	
New code	516.30	Idiopathic interstitial pneumonia, not otherwise specified Idiopathic fibrosing alveolitis	
New code	516.31	Idiopathic pulmonary fibrosis cryptogenic fibrosing alveolitis	

Nonspecific Interstitial Pneumonitis

Nonspecific interstitial pneumonitis (NSIP) is one of the idiopathic interstitial pneumonias, a group of scarring lung diseases with distinctive presentations, pathophysiology and clinical course. NSIP includes a group of interstitial lung disorders with a more favorable prognosis that need to be distinguished from idiopathic pulmonary fibrosis (IPF), and other idiopathic interstitial pneumonias. The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for NSIP, as well as for a number of other interstitial lung diseases.

Onset of NSIP is often between ages 40-50 yrs, thus about a decade or more younger than patients with IPF. Unlike IPF, NSIP may occur in children. Onset is usually gradual, but may be subacute. The duration of symptoms before diagnosis may range from 6 months to 3 years. Breathlessness, cough, and fatigue are usual symptoms, and many have weight loss. Lung function tests show similar but milder physiological abnormalities than those found in IPF; that is, a restrictive ventilatory defect in more than 90% of patients, and mild airflow limitation in a minority. Most develop hypoxemia during exercise. The NSIP histological pattern encompasses a broad spectrum of histologic features with varying degrees of alveolar wall inflammation or fibrosis.

Nonspecific interstitial pneumonitis is also called nonspecific interstitial pneumonia. While this is not indexed in ICD-9-CM, interstitial pneumonia is indexed to 516.8, Other specified alveolar and parietoalveolar pneumonopathies. For NSIP as an idiopathic interstitial pneumonia, ATS-ACCP would currently use code 516.3, Idiopathic fibrosing alveolitis. The NSIP histologic pattern may also be associated with other clinical causes, including collagen vascular disease, hypersensitivity pneumonitis, drug-induced pneumonitis, infection, and immunodeficiency. If it is due to some other underlying cause, then code 516.8 should be used.

Idiopathic NSIP has a different course, and a more favorable prognosis than IPF, and thus needs to be distinguished from IPF, and other idiopathic interstitial pneumonias. A specific code will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References

Martinez FJ. Idiopathic interstitial pneumonias: usual interstitial pneumonia versus nonspecific interstitial pneumonia. Proc Am Thorac Soc. 2006;3(1):81-95. (Available here: http://pats.atsjournals.org/cgi/content/full/3/1/81).

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise

516.3 Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia

New code

516.32 Idiopathic non-specific interstitial pneumonitis

Excludes: Non-specific interstitial pneumonia NOS, or due to known underlying cause (516.8)

516.8 Other specified alveolar and parietoalveolar pneumonopathies

Add

Excludes: Idiopathic non-specific interstitial pneumonitis (516.32)

Acute Interstitial Pneumonia

Acute interstitial pneumonia (AIP) is a rapidly progressive and histologically distinct form of interstitial pneumonia. The histopathology is described as an organizing form of diffuse alveolar damage identical to the histologic pattern found in acute respiratory distress syndrome (ARDS) caused by sepsis and shock. Some of the cases described by Hamman and Rich probably represented AIP. The term AIP is reserved for cases of unknown cause.

AIP occurs over a wide age range, with a mean age of approximately 50 yr. Patients often have a prior illness suggestive of a viral upper respiratory infection with constitutional symptoms such as myalgias, arthralgias, fever, chills, and malaise. There is no proven treatment, and mortality rates are high (50% or more). Most deaths occur between 1 and 2 mo. of illness onset.

The term “acute interstitial pneumonia” is indexed in ICD-9-CM to code 136.3, Pneumocystosis. The histologic pattern in AIP of diffuse alveolar damage may also occur due to Pneumocystis carinii pneumonia, as well as due to cytomegalovirus, due to collagen vascular disease, due to a drug-induced pneumonitis, due to ARDS in sepsis or shock, or due to a number of other potential causes. However, at this time, based on the 2001 consensus s statement of the American Thoracic Society and the European Respiratory Society, the term AIP is reserved for cases of unknown cause, thus represents a specific idiopathic interstitial pneumonia.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for AIP, as well as for a number of other interstitial lung diseases. There is no specific ICD-9-CM code for AIP. ATS-ACCP stated most clinicians would currently code AIP to code 516.3, Idiopathic fibrosing alveolitis, although code 516.8, Other specified alveolar and parietoalveolar pneumonopathies, might also be used. AIP has a different course, and a less favorable prognosis than other idiopathic interstitial pneumonias, and thus needs to be distinguished from them. A specific code will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References

Bouros D, Nicholson AC, Polychronopoulos V, du Bois RM. Acute interstitial pneumonia. Eur Respir J. 2000 Feb;15(2):412-8.
(Available here: http://erj.ersjournals.com/cgi/reprint/15/2/412.)

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise 516.3 Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia

New code 516.33 Acute interstitial pneumonia

Excludes: Pneumocystis pneumonia (136.3)

Respiratory Bronchiolitis-associated Interstitial Lung Disease

Respiratory bronchiolitis-associated interstitial lung disease (RB-ILD is the clinical manifestation of interstitial lung disease associated with the pathologic lesion of respiratory bronchiolitis. Respiratory bronchiolitis is a histopathologic lesion found in cigarette smokers and is characterized by the presence of pigmented intraluminal macrophages within respiratory bronchioles. It is rarely symptomatic and is usually associated with no more than minor small airway dysfunction. However, in rare cases the condition presents as a form of interstitial lung disease with significant pulmonary symptoms, abnormal pulmonary function, and imaging abnormalities. It is then described as respiratory bronchiolitis-associated interstitial lung disease (RB-ILD).

Nearly all patients with RB-ILD present with nonspecific respiratory complaints including gradual onset of dyspnea and the presence of a new or changed cough. It usually affects current smokers in the fourth and fifth decades of life with history of heavy smoking (usually more than 30 pack-years). Men are more often affected than women, by a ratio of almost 2:1. Due to the consistent relationship of RB-ILD with smoking, other disorders are commonly found with it , including centrilobular emphysema.

In respiratory bronchiolitis the histologic features include changes that are patchy at low magnification and have a bronchiolocentric distribution. Respiratory bronchioles, alveolar ducts, and peribronchiolar alveolar spaces contain clusters of dusty brown macrophages. Intraluminal macrophages are accompanied by a patchy submucosal and peribronchiolar infiltrate of lymphocytes and histiocytes. Mild peribronchiolar fibrosis is also seen and expands contiguous alveolar septa, which are lined by hyperplastic type II cells and cuboidal bronchiolar-type epithelium.

RB-ILD has been linked to desquamative interstitial pneumonia (DIP). DIP is considered to be a more extensive form of RB-ILD in which the pigmented macrophages fill alveolar spaces diffusely throughout larger areas of the lung. RB-ILD may be regarded as on a spectrum with DIP, depending on the extensiveness of the alveolar macrophage accumulation, although there are differences in the clinical presentation, imaging findings, and prognosis between the two patterns and as such they are described separately.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for RB-ILD, as well as for a number of other interstitial lung diseases. There is no specific ICD-9-CM code for respiratory bronchiolitis interstitial lung disease; the term is not indexed in ICD-9-CM. The term interstitial lung disease is indexed to code 515, Postinflammatory pulmonary fibrosis. ATS-ACCP stated most clinicians would currently code RB-ILD to code 516.8, Other specified alveolar and parietoalveolar pneumonopathies. RB-ILD has a different course, and a more favorable prognosis than many other idiopathic interstitial pneumonias, and thus needs to be distinguished from them. A specific code will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References

Portnoy J, Veraldi KL, Schwarz MI, Cool CD, Curran-Everett D, Cherniack RM, King TE Jr, Brown KK. Respiratory bronchiolitis-interstitial lung disease: long-term outcome. Chest. 2007 Mar;131(3):664-71. PubMed PMID: 17356078. (http://chestjournal.chestpubs.org/content/131/3/664.full.pdf+html)

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise	516.3	Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia
New code	516.34	Respiratory bronchiolitis interstitial lung disease

Lymphocytic Interstitial Pneumonia

Lymphocytic interstitial pneumonia (LIP), or lymphoid interstitial pneumonia, is characterized by the infiltration of the pulmonary interstitium with lymphocytes and plasma cells. The position of LIP within classification systems has changed with advances in understanding the nature of pulmonary lymphoid infiltrates, and LIP was previously classified under the heading of pulmonary lymphoproliferative disorders by many groups, with the idea that many cases would progress to lymphoma. However, with the advent of immunohistochemistry and molecular analysis, reactive and neoplastic infiltrates are usually separable, with only a small number of cases of LIP found to actually undergo malignant transformation. With regard to histogenesis, LIP is perhaps best regarded as a histologic variant of diffuse pulmonary lymphoid hyperplasia with predominantly interstitial changes. A related condition, follicular bronchiolitis, is predominantly a peribronchial (or peribronchiolar) lymphocytic infiltrate with germinal centers.

Onset of LIP is often slow with gradually increasing cough and breathlessness over 3 or more years. Fever, weight loss, chest pain, and arthralgia are occasionally found. LIP is more common in women. Although it may present at any age, it is most typically diagnosed in the fifth decade.

Lung nodules and widespread consolidation may occur. LIP is defined as a dense interstitial lymphoid infiltrate, including lymphocytes, plasma cells, and histiocytes with associated Type II cell hyperplasia and a mild increase in alveolar macrophages. The alveolar septa should be extensively infiltrated. Lymphoid follicles, including follicles with germinal centers, are often present, usually in the distribution of pulmonary lymphatics. More than one-third progress to diffuse fibrosis, and it is unclear whether treatment with corticosteroids influences the course of the disease or has a significant effect on lung physiology. Occasional cases resolve or improve substantially.

The LIP pattern may be caused by a number of clinical conditions. There are cases of idiopathic LIP, where the cause is unknown. However, cases of LIP must be thoroughly investigated clinically for any known cause or associations, such as collagen vascular diseases, immunodeficiency, infection such as Pneumocystis carinii pneumonia, drug-induced or toxin exposure, and autoimmune diseases.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for lymphocytic interstitial pneumonia, as well as for a number of other interstitial lung diseases, with LIP to be included with the idiopathic interstitial pneumonias. There is currently no specific ICD-9-CM code for lymphocytic interstitial pneumonia or lymphoid interstitial pneumonia. However, lymphoid interstitial pneumonia is included at code 516.8, Other specified alveolar and parietoalveolar pneumonopathies.

This proposal would create a new specific code for idiopathic lymphoid interstitial pneumonia, at subcategory 516.3, with other idiopathic interstitial pneumonias. However, it would exclude lymphoid interstitial pneumonia NOS or due to known cause to 516.8, Other specified alveolar and parietoalveolar pneumonopathies. Additional notes would also be created. A specific code for idiopathic LIP will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References:

Guinee DG Jr. Update on nonneoplastic pulmonary lymphoproliferative disorders and related entities. Arch Pathol Lab Med. 2010 May;134(5):691-701. Review. PubMed PMID: 20441500
(Available here: http://www.archivesofpathology.org/doi/pdf/10.1043/1543-2165-134.5.691.)

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise — 516.3 Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia

New code — 516.35 Idiopathic lymphoid interstitial pneumonia
Idiopathic lymphocytic interstitial pneumonitis

Excludes: Lymphoid interstitial pneumonia NOS, or due to known underlying cause (516.8)
Pneumocystis pneumonia (136.3)

516.8 Other specified alveolar and parietoalveolar pneumonopathies

Add — Code first, if applicable, underlying cause of pneumonopathy, if known

Add — Use additional E code, if applicable, for drug-induced or toxic pneumonopathy

Add — Excludes: Idiopathic lymphoid interstitial pneumonia (516.35)

Cryptogenic Organizing Pneumonia

Cryptogenic organizing pneumonitis, or cryptogenic organizing pneumonia, is a clinicopathologic entity involving a histopathologic organizing pneumonia of unknown cause. The term cryptogenic organizing pneumonitis (COP) is preferred over prior names such as bronchiolitis obliterans with organizing pneumonia (BOOP), because it conveys the essential features of the syndrome and avoids confusion with airway diseases such as constrictive bronchiolitis obliterans.

In COP, Patients typically present with an illness of relatively short duration (usually less than 3 mo) with variable degrees of cough and dyspnea. Lung function tests confirm a restrictive ventilatory pattern (usually mild to moderate), with a moderately reduced diffusing capacity in most. Airflow obstruction is present in a minority, but is thought to be an independent consequence of smoking. Mild resting hypoxemia may be present and reflects marked disturbance of gas exchange. There is an equal sex distribution for COP, but nonsmokers outnumber smokers by 2:1. Mean age of onset is 55 yr.

Histologically, the organizing pneumonia pattern is a patchy process characterized primarily by organizing pneumonia involving alveolar ducts and alveoli with or without bronchiolar intraluminal polyps.

The majority of patients with COP recover completely on administration of oral corticosteroids, but a significant number relapse within 1 to 3 months when the corticosteroids are reduced or stopped. Prolonged treatment for 6 months or longer is advised.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for COP, as well as for a number of other interstitial lung diseases. There is currently no specific ICD-9-CM code for COP, but it is indexed to code 516.8, Other specified alveolar and parietoalveolar pneumonopathies. Since it is idiopathic, some may also use code 516.3, idiopathic fibrosing alveolitis, according to ATS-ACCP.

This proposal would create a new specific code for COP, at subcategory 516.3, with other idiopathic interstitial pneumonias. However, it would exclude organizing pneumonia NOS or due to known cause to 516.8, Other specified alveolar and parietoalveolar pneumonopathies. A specific code for COP will facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References

Cordier JF. Cryptogenic organizing pneumonia. Eur Respir J. 2006 Aug;28(2):422-46. Review. PubMed PMID: 16880372. (Available here: http://erj.ersjournals.com/cgi/content/full/28/2/422)

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise

516.3 Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia

New code

516.36 Cryptogenic organizing pneumonia

Excludes: organizing pneumonia NOS, or due to known underlying cause (516.8)

516.8 Other specified alveolar and parietoalveolar pneumonopathies

Add

Excludes: cryptogenic organizing pneumonia (516.36)

Desquamative Interstitial Pneumonia

Desquamative Interstitial Pneumonia (DIP) involves accumulation of macrophages in alveoli. The disorder was originally thought to represent desquamation of epithelial cells. Another term considered for it is alveolar macrophage pneumonia. It affects primarily cigarette smokers in their fourth or fifth decades of life. DIP is more common in men than in women by a ratio of 2:1. Insidious onset of dyspnea and dry cough over weeks or months is usual and patients may progress to respiratory failure.

Lung function tests show normal lung volumes or a mild restrictive abnormality, and moderately decreased diffusing capacity. The prognosis of DIP is generally good, as most patients improve with smoking cessation and corticosteroids. The overall survival is about 70% after 10 yr.

The histologic DIP pattern is characterized by diffuse involvement of the lung by numerous macrophage accumulations within most of the distal airspaces. The alveolar septa are thickened by a sparse inflammatory infiltrate that often includes plasma cells and occasional eosinophils, and they are lined by plump cuboidal pneumocytes. Lymphoid aggregates may be present. The main feature that distinguishes DIP from respiratory bronchiolitis (RB) interstitial lung disease (ILD) is that DIP affects the lung in a uniform diffuse manner and lacks the bronchiolocentric distribution seen in RB. The intraluminal macrophages in DIP frequently contain dusty brown pigment identical to that seen in RB. Finely granular iron may be seen in the macrophage cytoplasm.

The American Thoracic Society (ATS) and the American College of Chest Physicians (ACCP) have requested creation of a specific code for DIP, as well as for a number of other interstitial lung diseases. There is currently no specific ICD-9-CM code for COP, but it is included at code 516.8, Other specified alveolar and parietoalveolar pneumonopathies. Since it is idiopathic, ATS-ACCP proposes that it be classified at subcategory 516.3, with other idiopathic interstitial pneumonias. A specific code for DIP will enable differentiation from RB-ILD and other interstitial lung disease, and facilitate epidemiological, clinical, comparative effectiveness and cost effectiveness research.

References

Ryu JH, Myers JL, Capizzi SA, Douglas WW, Vassallo R, Decker PA. Desquamative interstitial pneumonia and respiratory bronchiolitis-associated interstitial lung disease. Chest. 2005 Jan;127(1):178-84. PubMed PMID: 15653981. (Available here: http://chestjournal.chestpubs.org/content/127/1/178.full.pdf+html.)

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TABULAR MODIFICATIONS

516 Other alveolar and parietoalveolar pneumonopathy

Revise	516.3	Idiopathic ~~fibrosing alveolitis~~ interstitial pneumonia
New code	516.37	Desquamative interstitial pneumonia